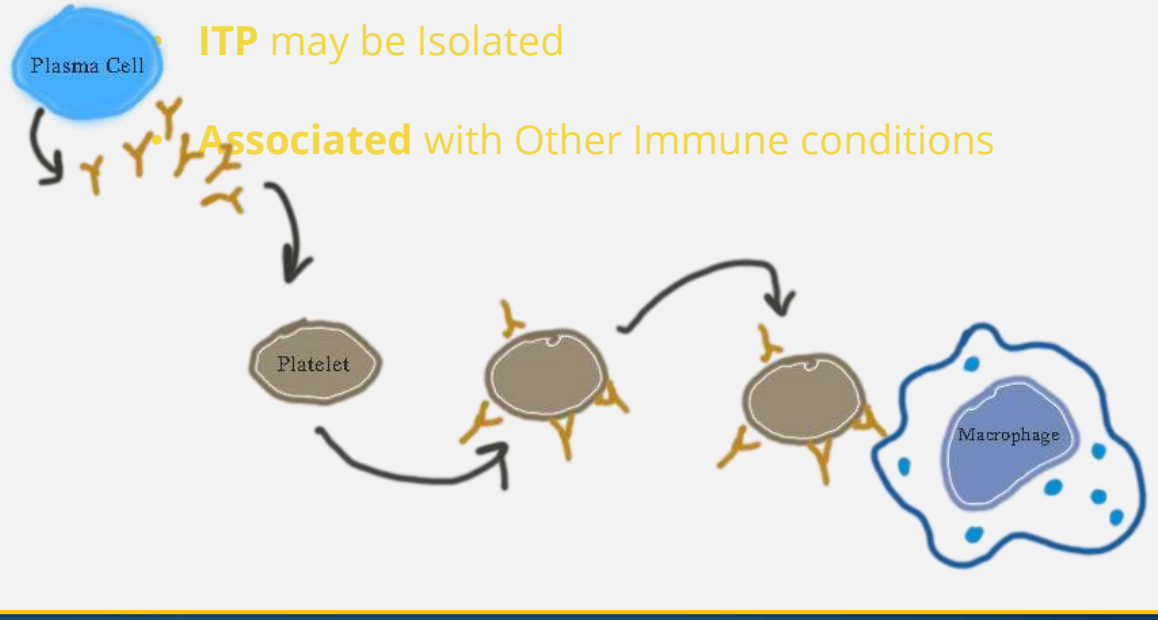
Idiopathic Thrombocytopenic Purpura

(ITP)

Idiopathic Thrombocytopenic Purpura

- **Immune-mediated** Disease
- **Antiplatelet Antibodies**
  - Against **Glycoprotein IIb/IIIa** on membrane

Ab attach to Gp IIb/IIIa on Platelets → Sensitize **Platelets** → Recognition by Reticuloendothelial System Cells → Removal from circulation



- **ITP** may be Isolated
- **Associated** with Other Immune conditions

Clinical Features & Diagnosis

Clinical **Features**

- **Acute & Self-limiting** in Children – Preceded usually by viral infection 2 weeks before ITP
- **Adults**
  - **Females** affected more
  - **Insidious** onset
  - Chronic, remitting relapsing course
  - *No preceding history* of viral infection
- Presentation depends on degree of Thrombocytopenia

Platelets

- **>50 x 10^9/L** – Accidentally discovered
- **<20 x 10^9/L** – Spontaneous Bleeding
- **>20 x 10^9/L** – Easy bruising
  - Epistaxis
  - Menorrhagia

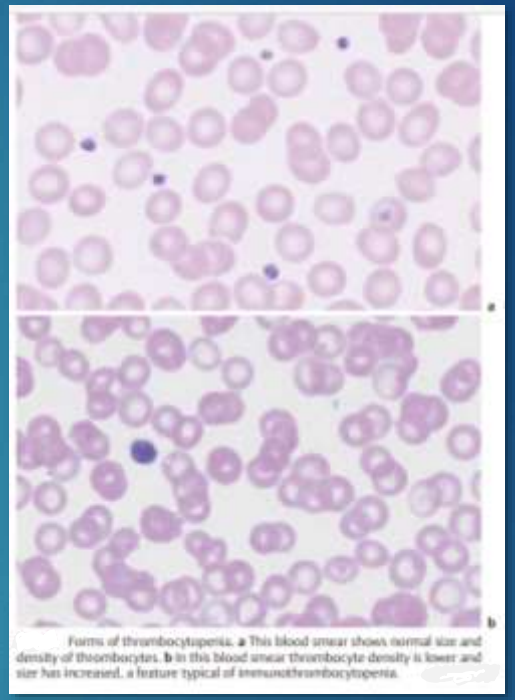
- Symptoms & Signs of Connective tissue disease

Clinical Pearl

There is no splenomegaly in ITP

Investigations

- **Complete Blood Counts -** Thrombocytopenia
- **Peripheral Blood Film -** Reduced platelet number
- **Antiplatelet antibodies -** Present
- **Bone marrow biopsy –**
  - Usually not necessary for the diagnosis
  - Increased Megakaryocytes
  - Do in > 65 years old to look for B-cell malignancy
- **Autoantibody testing** for Connective Tissue Disease
- **HIV** Testing



Management

Treatment

- **Stable compensated ITP & Platelets >30×10^9 /L** - ***No Treatment*** *except* at times of increased bleeding risk e.g., as surgery & biopsy

First Line Therapies

1. **Glucocorticoids**
2. **Intravenous Immunoglobulins (IVIG)**
3. **Platelet transfusions**

- ITP may become chronic, with remissions & relapses
- Relapses treated by Glucocorticoids

Glucocorticoids

- First-line therapy
- Spontaneous bleeding *or* Platelets < 20 x 10^9/L
- High doses, either Prednisolone (1 mg/kg daily) or Dexamethasone (40 mg daily for 4 days)
- Pulsed Methyprednisolone may also be used
- Suppresses antibody production & inhibit phagocytosis of sensitised platelets by reticulo-endothelial cells
- Aim: Platelets >30 x 10^9/L
- Reduce the dose of steroids after remission

IVIG

- Block antibody receptors on reticuloendothelial cells & inhibiting platelet clearance by these
- Combined with glucocorticoid therapy if there is significant mucosal bleeding, *or* a slow response to glucocorticoids alone

Platelet Transfusion

- If persistent *or* potentially life-threatening bleeding or during splenectomy

Treatment

Second Line Therapies

- **Indications -** 2 Relapses *or* Primary Refractory Disease
- **Options**
  - *Immunosuppression*
  - *Splenectomy*
  - *Thrombopoietin Receptor Agonists (TPO-RA)*
- The order in which therapies should be used is not clear

Immunosuppression

- Low-dose glucocorticoid therapy & Immunosuppressants *(Rituximab, Ciclosporin, Mycophenolate & Tacrolimus)* - Produce Remissions & Reduce Relapses

Splenectomy

- Complete remission in ~ 70% patients & improvement in a further 20–25% in favorable cases
- Precautions for splenectomy

TPO-RAs

- *Eltrombopag – Oral TPO-RA*
- *Romiplostim – Injectable TPO analogue*
- Response in around 75% of cases

Splenectomy Patient Care

i. Vaccinate with Pneumococcal, Haemophilus influenzae Type B, Meningococcal group C & Influenza vaccines at least 2–3 weeks before elective splenectomy. Vaccination should be given after emergency surgery but may be less effective

ii. Pneumococcal booster dose should be given at least 5-yearly & Influenza annually. Vaccination status must be documented.

iii. Life-long prophylactic Penicillin-V is recommended. Consider a Macrolide in penicillin-allergic patients.

iv. Patients should be educated regarding the risks of infection & methods of prophylaxis.

v. Card *or* Bracelet alert should be carried to alert health professionals to the risk of overwhelming sepsis.

vi. In sepsis, patients should be resuscitated, and given IV antibiotics to cover Pneumococcus, Haemophilus and Meningococcus, according to local resistance patterns.

vii. The risk of cerebral malaria is increased in the event of infection.

viii. Animal bites should be promptly treated with local disinfection and antibiotics, to prevent serious soft tissue infection & sepsis.

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